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Vulvar intravascular papillary endothelial hyperplasia or Masson's tumor: A case report

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ABSTRACT

Masson's tumor, also known as intravascular papillary endothelial hyperplasia (IPEH), is a rare and benign vascular disease in which there is a reactive hyperplasia of intravascular endothelial cells. This tumor is most commonly found in soft tissues in the head, neck and upper extremities. We report a rare case of IPEH on the vulva. A Hispanic woman in her mid-thirties presented with a painful and pruritic left vulvar mass. On physical examination, a pedunculated mass was visualized on the left labia majora. Under pathologic examination, it was concluded the lesion was IPEH and it was surgically excised. This is a rare case of IPEH located on the vulva. However, despite this rarity, a simple local excision could be used to manage IPEH.

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1. Introduction

Intravascular papillary endothelial hyperplasia, also known as Masson's tumor, is a rare tumor that affects intravascular endothelial cells [1]. It is usually found in soft tissues of the head, neck and upper extremities [2]. It is best treated with complete surgical resection [3]. We report a case of intravascular papillary endothelial hyperplasia of the vulva which was successfully managed by complete surgical excision.

2. Case Presentation

A Hispanic woman in her mid-thirties, gravida 0, with a history of bipolar disorder, presented with a chief complaint of a left vulvar mass for one year. She described the lesion as "purplish" and complained of tenderness to the touch. The mass had increased in size over the course of the year along with worsening pain.

Previous evaluation ruled out herpes simplex virus and the leading differential diagnosis was vulvar varicosities based on the appearance and characteristics on examination. She was treated accordingly with conservative measures and topical hydrocortisone cream for the discomfort.

Due to increasing size and pain, however, she sought re-evaluation.

Her medical history was non-contributory, and she was up to date on recommended health maintenance evaluations.

On examination, a pedunculated mass was identified on the left labia majora (Fig. 1). It was grayish to tan brown in color, and it measured approximately 2 cm × 3 cm. The mass surface had exposed, granulation tissue and a well demarcated outline near the base. The surface appeared irregular and wrinkled. Central areas of hemorrhagic tissue necrosis were also noted. The mass and base had minimal tenderness to palpation. Tender lymph nodes were identified on the left groin, with no evidence of excoriations. Vaginal exam demonstrated copious white discharge, without a foul smell. The rest of the pelvic examination did not reveal any related abnormalities.

Clinical biopsy revealed granulation tissue with acute and chronic inflammation. Additional testing was negative for human papillomavirus 16 and 18 and fungi by special stain. Vaginal discharge was collected and tested positive for bacterial vaginosis. She was prescribed metronidazole. Blood samples were taken and tested for syphilis and human immunodeficiency virus and results were negative. The mass was excised in the operating room.

Under pathologic examination, a well-circumscribed lesion, with characteristic papillary fronds, was visualized. The proliferative process was entirely limited to the intravascular spaces. The papillae were composed of fibrohyalinized tissue of two or more endothelial cell layers. Individual endothelial cells showed mild hyperchromasia and mild nuclear atypia; however, mitotic figures were not easily identified. It was concluded that the lesion was intravascular papillary endothelial

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Fig. 1. Gross images of vulvar lesion.

hyperplasia (Fig. 2). The patient recovered uneventfully without evidence of recurrence.

3. Discussion

Masson's tumor was first discovered by Pierre Masson in 1923. Masson initially named the tumor vegetant intravascular hemangioendothelioma and described it as a neoplasm [1]. In 1929, Folke Henschen re-described it as a reactive process instead of a neoplasm and named the lesion endovasculite proliférante thrombopoïétique [4]. Later, in 1976, the lesion was renamed intravascular papillary endothelial hyperplasia (IPEH) by Kevin Clearkin and Dr. Franz Enzinger [5]. IPEH, or Masson's tumor, is a rare and benign vascular disease in which there is a reactive hyperplasia of intravascular endothelial cells [1]. It consists of an intravascular proliferation of numerous papillae with a core of connective tissue and an endothelial surface [6]. This tumor is most commonly found in soft tissues in the head, neck and upper extremities, but it can affect any part of the body [2].

Masson's tumor can be classified into three types: the primary or pure form, the secondary or mixed form, and the extravascular form [7]. The primary/pure form, which is the most common form, typically occurs in a dilated vessel, most usually a vein rather than an artery,

and arises in subcutaneous soft tissue [8]. The secondary/mixed presents in preexisting vascular abnormalities. Finally, the extravascular form, which is the least common form, occurs in hematomas [9].

Masson's tumor can be mistaken for other subcutaneous tumors, such as lipomas, angiomas, vascular malformations, and cysts. However, it most commonly resembles, and can be mistaken for, an angiosarcoma [10]. It is important to differentiate between IPEH and these other conditions. This is done through a clinical and histological diagnostic work-up [11]. Clinically, Masson's tumor will appear as a firm or soft surface nodule, blue-red in color, and non-pulsatile [11]. Histologically, it will be characterized by proliferating endothelium lining papillary and anastomosing channel-like structures [12]. In addition, it will not have any features of malignancy, such as necrosis, cellular atypia, and mitotic figures, which helps to distinguish it from the similar angiosarcoma [10].

Immunohistochemical confirmation may be required only if the endothelial origin of the lesion is in question [13]. In such cases, endothelial cell markers, such as von Willebrand factor, CD31, factor XIIIa, and CD43, may be used, which would highlight the endothelial lining around the papillary tufts [13].

There are many theories regarding Masson's tumor's cause and pathogenesis, but ultimately it is unknown [14]. Risk factors include history of local trauma or previous vascular conditions (i.e. hemangiomas,

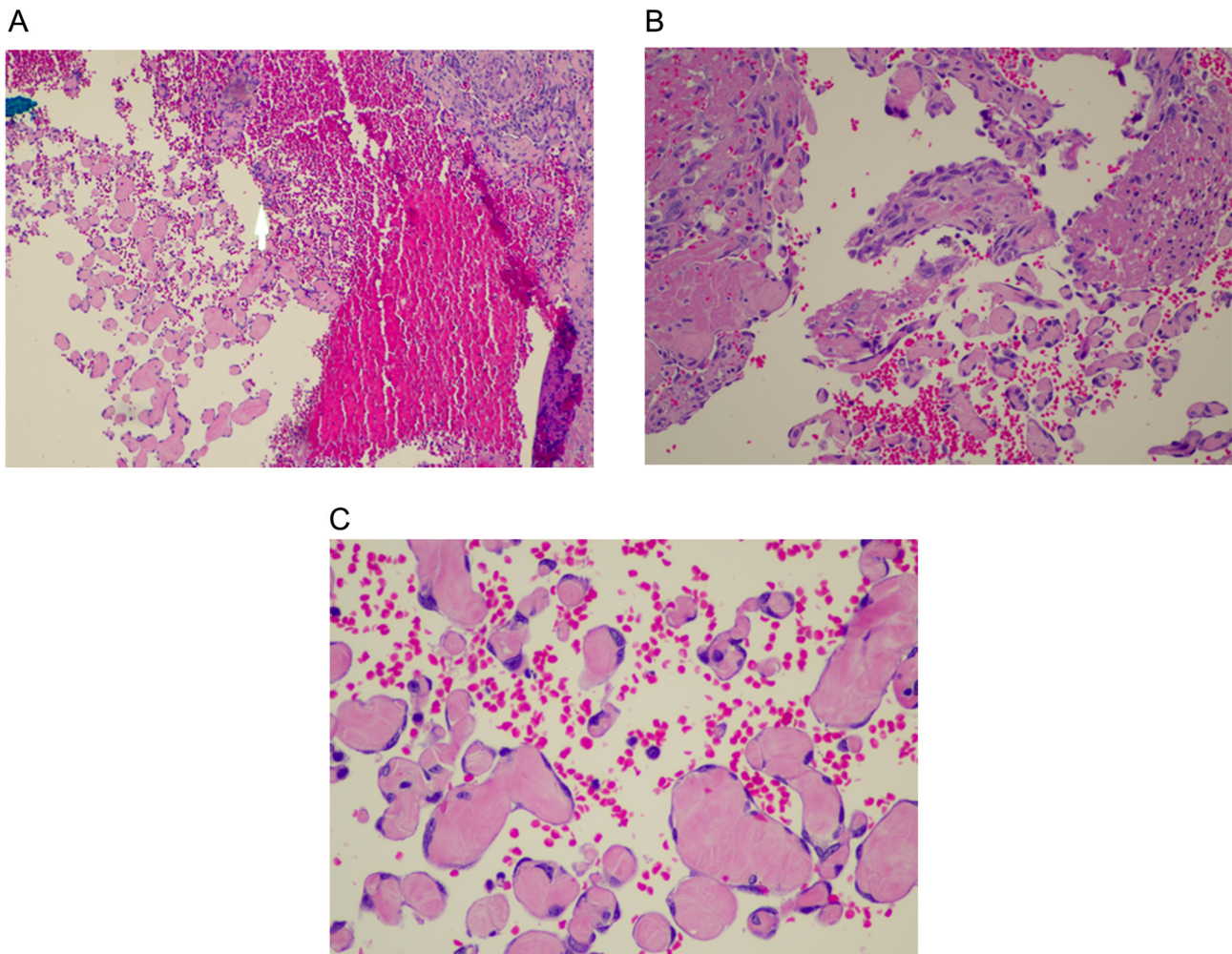


Fig. 2. Histopathology images of the dissected lesion. A. in 10× - Sections show dilated vessel containing papillary proliferation of plump endothelial cells without atypia. B. in 20× - Fibrin deposition and thrombi are present. C. in 40× - There is variable nuclear pleomorphism. No mitotic figures, no necrosis, no solid cellular areas are present.

blood stasis, etc.). Most of the time it will be idiopathic [14]. Women are at a higher risk than men of acquiring Masson's tumor¹⁵. It can affect all age groups and has no race or ethnic group preferences [15].

Treatment of Masson's tumor consists of complete surgical resection in order to decrease the risk of recurrence [3]. It has a good prognosis, with no evidence of local invasion or metastasis [16].

This is a rare case of a patient with intravascular papillary endothelial hyperplasia, also known as Masson's tumor, located on the vulva. IPEH is typically found in the head, neck and upper extremities. It can be found in other places on the body, but it is rare to find in the genital region. To our knowledge, there is only one case of IPEH located on the vulva, which was described by Beutler and Cohen in 2016 [17]. Beutler and Cohen described IPEH as an organizing thrombus in the vascular lumen with accompanying hyperplastic endothelial cell proliferation and reiterated the rarity of Masson's tumor being located on the vulva, as opposed to the typical areas, such as the head and the limbs. However, despite this rarity, a simple local excision can always be used to manage Masson's tumor [17].

4. Conclusion

The importance of IPEH lies in the fact that it histologically simulates angiosarcoma. The lack of cytologic atypia as well as lack of necrosis help to differentiate it from angiosarcoma. Moreover, IPEH tends to

recur if incompletely resected. Correct diagnosis of this entity is essential to prevent aggressive treatment.

Contributors

Vania Nwokolo helped to obtain consent of the patient, and helped to write up the case report.

Ravindra Veeramachaneni helped to write the case report and provided and described the histopathological images.

Saul Rivas helped to write the case report and obtained the gross images.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient consent

Obtained.

Provenance and peer review

This case report was peer reviewed.

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